SCLERODERMIA AND MYOSITIS

BY

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SCLERODERMIA AND MYOSITIS.[1]

SOME three years ago I contributed to the *Bristol Medico-Chirurgical Journal*[2] an account of a case of muscular atrophy with sclerodermia, in which the evidence that the changes in the muscle were primary—either of the nature of a progressive atrophy or a dystrophy—was irresistible. Since publishing that case four other patients have come under my observation at the Bristol Royal Infirmary who seem to provide material for a further study of these conditions and to throw fresh light on their nature. Of recent years, too, other cases have been recorded which make it evident that the associated affections met with in sclerodermia are no longer the isolated conundrums which they formerly seemed. From a careful clinical investigation of the cases under my own care and from the published records of other observers I think that there is beginning to be collected a considerable amount of evidence that the muscular changes of sclerodermia are (together with the skin changes themselves) merely a local manifestation of a general disease, an infective inflammatory affection resembling in many points the rheumatic infection.

Just as in acute rheumatism we are familiar with the usual form of a general shifting synovitis, varied at times by an attack on serous and especially cardiac membranes, manifesting itself sometimes by localised tendinous infiltrations of a nodular type, at others by subcutaneous erythematous inflammation, or again appearing in cerebral form as chorea,

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[1] A paper read at a meeting of the Bristol Medico-Chirurgical Society on Nov. 14th, 1906.

[2] Bristol Medico-Chirurgical Journal, December, 1903.

so there is evidence of an analogous polymorphism in the disease which for convenience may he termed generally sclerodermia. There is little need to describe the clinical appearances of sclerodermia, the localised form with its patch of glossy, hard, ivory skin, or the generalised form with more or less widespread "hidebound" appearance, thick, immobile, parchment integument, with the natural folds of the skin obliterated, and expression lost in the face giving rise to the "masque sclérodermique."

Of the pathology much has been written and little explained. Briefly, the cellular elements of the skin disappear, together with the elastic and fatty tissue, the vessels become narrowed or obliterated, and the collagenous bundles hypertrophy. Ultimately the whole affected area becomes atrophied into a thin parchment-like cutaneous plate without a papillary layer or subcutaneous tissue but covered with atrophied epidermis.[3] Nerve lesions have been reported but the evidence of any constant nervous origin for this affection is still lacking. In such cases as have been examined after death the muscle changes have been variously reported to have been either of an inflammatory nature, "myositis diffusa," or, in what may be interpreted to be a later stage, of the nature of a fibrous degeneration or infiltration, "myositis fibrosa." It must be confessed that the necropsies on record throw little or no light so far on the etiology of the disease. We are thrown back again on the living subject, to an investigation of the clinical phenomena, to an examination of analogous diseases and allied conditions, and perhaps to a research into the manner of discharging the vital functions. Bloch and Retmann[4] have recently published their researches upon metabolism in sclerodermia and state that there is no characteristic variation in the Nitrogen-exchange. By correlation of these one with another and by comparison or contrast with the processes observed in other diseases it may be possible to arrive at some definite conclusion as to what sclerodermia is and how it arises. Although the four cases described may be quite wrongly associated, although the

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[3] Fibromatosis or new formation of collagen fibres takes place not only in association with granulating wounds but also in a large number of affections of the corium. After recurrent attacks of lymphangitis and erysipelas where a persistent œdema has occurred the œdema causes a rarefaction of the fibrous stroma, which is followed by a fibromatous process. In sclerodermia the hardness is due to a hypertrophy of the fibrous tissue. Pathology of Skin Disease, Macleod, p. 185.

[4] Bloch and Retmann: Wiener Klinische Wochenschrift, vol. xix., p. 630.

suggested points of similarity may be quite wrongly interpreted, still there remain some features in common which bespeak the same pathological processes underlying them.

CASE 1.—A healthy girl, aged 13 years, first came under the care of Dr. H. Waldo 18 months ago on account of a patch of painful, thickened, stiff, glossy skin on the right shoulder and arm, a narrow band extending from the posterior aspect of the acromion over the deltoid and down the outer and posterior part of the arm to a point a few inches below the elbow. It did not at first occupy so large an area as this but spread rapidly in the first week or so, then became stationary, and finally began to disappear, especially behind the shoulder where now the skin is practically normal; the white waxy appearance showed itself almost at once. Three months ago she developed a similar smaller patch about two inches long and one inch wide, oval in shape, on the left thigh just over the apex of Scarpa's triangle, and now she is developing some indefinite thickening of the skin on the inside of the left knee just above the condyle of the femur. Also within a month or two of first attending the right hand was noticed to be smaller than the left, while the middle finger, although freely moveable as regards the joint, was incapable of being completely extended unless the wrist was flexed. The muscles of the forearm were not noticeably wasted nor lacking in power; flexion and extension of the fingers were perfectly normal up to a point where the tendon of the flexor sublimis digitorum to the middle finger seemed to be stretched to its utmost and the finger could be no further extended, the flexion only being apparent at the metacarpo-phalangeal joint. This is the condition at present: there are no involvement of tendon-sheath or fascia as in Dupuytren's contraction and no true primary muscular atrophy, all electrical reactions are normal, and it is neither a spastic contraction of the flexor muscle nor due to defective power in the extensor. This seems to be an essential shortening of muscle and tendon. The girl herself was not aware of any abnormality in this hand. There was no acute onset, no rheumatism, no pain, no tenderness, anæsthesia, or any symptom to attract the patient's attention.

CASE 2.—The patient was a single woman, aged 40 years, in domestic service. She showed changes in the skin of her left arm and hand which dated either from birth or very early infancy. She had true sclerodactylia, the thumb and

first finger of the left hand were deformed, the skin was thickened, hard, and cold but not white, and even the bones were irregularly thickened as if by periostitis. The skin was chiefly involved up the radial border of the forearm to a point just above the elbow so that on straightening the arm a thick fold of infiltrated skin was made tense and prominent, yet there was no implication of muscle or tendon in this part; but on the palmar aspect of the first metacarpal, where the skin was scarcely at all affected, the tendon of the flexor sublimis showed the characteristic contracture; the skin, fascia, and tendon sheath were all free from induration, yet the tendon was distinctly shortened, and on straightening the finger with the wrist and elbow extended it stood out as a thin cord tightly stretched. Quite recently this patient had severe pain and tenderness over the roots of the brachial plexus, but this had subsided without further change in skin or muscle.

CASE 3.—The patient was an unmarried woman, aged 22 years, the subject of a large goitre without symptoms of Graves's disease. Two years ago she was taken ill with an attack of rheumatism; not quite characteristic rheumatic fever, for there seemed to have been no effusion into the joints, only pain, severe pain, all over and inability to move the limbs. Recovering from this all her joints were found to be stiff and apparently enlarged, looking at first sight like osteo- or rheumatoid arthritis. But on examining the joints they were found to be limited in their movements, not by essential changes in the joints themselves, but by a general shortening of flexor tendons, of such a nature that if an attempt was made to straighten the various joints of the fingers, the wrist, and the elbow simultaneously the tendons became tense and complete extension was prevented; while flexion of the elbow immediately allowed the wrist to be extended. Further, the apparent expansion of the articular ends of the phalanges was shown by a radiogram not to depend upon bony changes but upon wasting of the soft parts. The muscles and tendons of the upper extremities alone showed these alterations and the electrical reactions were everywhere normal. In this patient the tendon sheaths and fascia were free and there was no sign of any skin affection. Although there was a goitre present without symptoms of Graves's disease, yet very small doses of thyroid extract or thyroideetin produced immediately tachycardia, nervousness, and tremor.

CASE 4.—The patient was a girl, aged 20 years, who previously was in good health and had suffered from no skin complaints. Two months ago she scratched her right arm. From this a general inflammation followed, apparently an ordinary attack of erysipelas, for which she was admitted to the Royal Infirmary. Now that the erysipelas had subsided there was over the insertion of the deltoid an induration of the skin about 6 inches long and 3 inches wide, roughly oblong in shape, slightly swollen, dusky red in colour but mottled with white streaks and stellate veins, surrounded by the "lilac ring." The edges were well defined but beyond the margin subcutaneous "islets" of infiltration could be felt which did not involve the epidermis. The skin did not feel adherent to the subjacent tissues, nor were any muscles affected. There was no pain and sensation was unaltered. The case seems to be one of localised sclerodermia or morphœa following directly upon erysipelas or starting from an infected wound. (At the present time this patient is suffering from an attack of articular rheumatism with acute endocarditis.)

The case which I reported three years ago is worth calling to mind in this connexion; to quote from the description then given, "The first symptom complained of was a weakness in the limbs; at first the left hand felt weak and numb, especially towards night ..... later the right hand followed suit and aching pains were constantly present in the forearms. Soon after, the legs began to ache on walking short distances so much that after a five mile round he was obliged to lie down and rest. ..... The tips of his fingers started tingling and were so sensitive that he gave up shoemaking, after vainly trying to relieve the tenderness by wearing woollen gloves." It was some months before any changes in the skin were noticed. Then the sclerodermia became gradually of the generalised type; there was atrophy of the muscles and marked contraction of some of them, notably the biceps, "which could be felt as a tense cord, no thicker than the little finger, lying under normal elastic skin which could easily be raised from the underlying muscle"; the right arm was flexed at an angle of nearly 90° at the elbow, while a lesser degree of wasting and contraction was shown in the left arm. Many other muscles had atrophied but none contracted like the biceps.

To Thibierge is due the credit of first seriously attracting attention to the changes in muscles associated with

sclerodermia[5] and more recently in *La Pratique Dermatologique*[6] he quotes Leredde and Thomas to show that the only well-established point in the pathogeny of the disease is that a sclerosis of the skin, possibly inflammatory in origin, is always present. A brief epitome of cases collected by Thibierge[7] is appended.

1. Girl, aged 17. Diffuse sclerodermia en plaques. Masque sclérodermique. Triceps wasted, movements free but feeble; arm can be flexed but not extended fully owing to contraction of biceps. General loss of power in hands and fingers, not depending on paralysis. Sterno-mastoids contracted. Peri-articular (scapulo-humeral) fibrous adhesions. R.D. in some muscles.

2. Woman, aged 23 (Westphal I.). Diffuse sclerodermia. Masque sclérodermique. Onset unnoticed. Left elbow cannot be extended owing to contraction of biceps which is as hard as wood. Hands and fingers half flexed in position of repose, and although the joints are relatively more freely moveable than usual flexion is very limited. No pain, some crepitation round joints, articular surfaces feel smooth. Leg muscles very hard, no true contraction, movements of knees limited. No paralysis.

Necropsy.—No joint changes. Fibrous peri-myositis, no interstitial myositis.

3. Woman, age 36 (Westphal II.). Diffuse sclerodermia. Masque sclérodermique. Many muscles atrophied although skin is free. Contraction in biceps similar to foregoing.

4. Woman, aged 53 (Goldschmidt). Sclerodactylia. Masque sclérodermique. Symmetrical gangrene. Skin of neck free. Muscles hard and leaden ("soudés") in the forearm; on movement dry leather creaking felt.

Necropsy.—Peri-muscular fascia very dense, inter-muscular planes of fascia show increase of fibrous tissue. Some atrophy of the muscle fibres.

5. Man, age 19 (Schultz). A fatal case of Addison's disease with sclerodermia and interstitial myositis.

Of other published cases I have been able to collect the following:—

6. Hutchinson.[8] A case of sclerodermia with contraction of sterno-mastoid not due to induration of the skin.

7. Grasset.[9] Sclerodermia with fibro-tendinous contractions due to a muscle atrophy, not myelopathic nor neuritic, but a simple "myopathy."

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[5] Thibierge: Revue de Médecine, 1890, vol. x., p. 291.
[6] Thibierge: La Pratique Dermatologique, Tome iv. Article: Sclerodermia. 1906.
[7] Revue de Médecine, 1890, vol. x., p. 291.
[8] Hutchinson: Archives of Surgery, 1891-92, vol. iii., p. 238.
[9] Grasset: Nouvelle Iconographie de la Salpêtrière, 1896, No. 5.

8. Elliott.[10] Amyotrophic changes preceding sclerodermia by five years.

9. Pelizaeus.[11] Sclerodermia with independent sclerosis of muscles.

10. Rosenfeld.[12] A similar case.

11. Haushalter and Spillman.[13] Girl, aged six. Diffuse sclerodermia en plaques accompanied by marked atrophy of muscles and arrested growth in one leg.

12. Herringham.[14] A man with sclerodermia and a condition of hands resembling rheumatoid arthritis but depending actually on muscular contractions, not joint changes; all the limbs contracted.

13. Fowle.[15] Diffuse sclerodermia with widespread tendon contractions.

14. Batten[16] in a very complete description of myositis fibrosa quotes a case of Liodner in which typical myositis fibrosa was associated with sclerosis o the skin.

15. Petjes and Clejat.[17] A fatal case of five years' duration in a woman who developed the most extensive sclerodermia and passed through a generalised acute myositis to a stage of fibrous contractions. Sclerodermia with myositis diffusa (necropsy).

Now the dependence of sclerodermia and the allied muscular atrophies upon nerve lesions is at best only a matter of conjecture; in the majority of necropsies no changes in the nervous system have been found; in the few instances on record where these changes did exist they were of such inconstant, even contradictory, types that no causal relationship could be established between nervous system, skin, and muscle. But the evidence of primary inflammation in the muscles of either an acute or chronic variety, ending as a rule in a fibrosis, is amply corroborated both by clinical and pathological facts. Beyond this it is difficult to penetrate clearly, the indications seem to point to an acute or chronic infection which affects mainly muscle and subcutaneous tissue, resulting in a sclerosis of the skin and the muscle, an infection to which perhaps persons suffering from other general diseases are peculiarly liable and especially the

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[10] Elliott: Journal of Cutaneous Diseases, May, 1897, p. 199.

[11] Pelizaeus: ref. Archives de Neurologie, 1897, No. 3, p. 139.

[12] Rosenfeld: Centralblatt für Neurologie, 1902, p. 976.

[13] Haushalter and Spillman: Nouvelle Iconographie de la Salpêtrière, 1899, xii., 200 (Obs. iii.).

[14] Herringham: Transactions of the Clinical Society, 1900, vol. xxxiii.

[15] Fowle: Boston Medical and Surgical Journal, 1904, vol. cli., p. 630.

[16] Batten: Transactions of the Clinical Society, 1904, vol. xxxvii., p. 12.

[17] Petjes and Clejat: Annales de Dermatologie et de Syphiligraphie, June, 1906.

subjects of Graves's disease, myxœdema, Raynaud's disease, Addison's disease, and nerve degenerations. It seems probable that myositis of a localised nature going on to sclerosis and contraction of a single muscle or isolated groups is commoner than generally supposed; and that the disease, of which this is only one manifestation, may run an acute, subacute, or chronic course, and on occasions may involve the skin, giving rise to one type of sclerodermia. A reference to the original monograph of Addison upon "Keloid of Alibert and True Keloid"[18] makes it obvious that the disease as seen and described by Addison under the latter title was not, as has always been represented, solely or even primarily a cutaneous disease, for he recognised and mentioned in two of his cases a general sclerosis of muscle, tendon, and fascia, together with—and as part of the same "morbid process very nearly allied to inflammation, probably of a strumous nature"—true keloid of the skin which we now call sclerodermia.

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[18] Addison: Opera Omn. New Sydenham Society, 1868. pp. 177 et seq.

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